Apoplexy in Young Adults—Diagnostic and Therapeutic Observations

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SUMMARY

Apoplexy in young adults is due to rupture of an artery or to obstruction, as by clot, by embolus, or by transitory vasospasm. In differential diagnosis, tumor and demyelinizing disease must be considered.

Spinal fluid study helps to differentiate between hemorrhagic stroke and stroke due to vascular block. In hemorrhagic stroke, search for the site of bleeding is imperative and angiography, the preferred method of study, should be carried out as soon as the patient's general condition permits.

In vascular occlusive stroke, measures to maintain the blood pressure and to dilate collateral channels are often helpful. Stellate block is most valuable in the acute phase immediately following embolism or thrombosis. In a small percentage of chronic cases, stellate block may be followed by significant recovery of function. Benefit may also be derived at times from systemic vasodilators such as niacin and intravenously administered histamine.

APOPLEXY, also referred to as "stroke," is the sudden diminution or paralysis of consciousness, sensation or voluntary motion resulting from the rupture or obstruction of an artery of the brain. The evidence of apoplexy may include any or all of the three objective factors either singly or in any combination.

Traditionally apoplexy has been regarded as one of the degenerative diseases affecting principally persons in the later years of life. Its occasional occurrence in young adults has been the object of intensified interest in recent years because of the development of improved modes of diagnosis and subsequent refinement of therapy. Young adults, by which is meant those persons who have not yet passed the fifth decade, may be subject to the same deteriorative diseases as are their elders. In addition, a considerable number of vasospastic diseases and embolic phenomena occur in this age group, and the likelihood of congenital anomalies, particularly saccular aneurysm and arteriovenous angioma, is greater. The general tissue reserve of young adults permits a wider range of therapeutic trial and provides a more substantial basis for objective evaluation of treatment.

The diagnosis of apoplexy in young adults, while generally simple, may on occasion be complicated. Other lesions capable of abrupt onset, such as brain tumor, or demyelinizing disease such as multiple sclerosis, may be initiated in a similar fashion. Differentiation between hemorrhagic stroke and that due to vascular blockage may also be complex, although as a rule, the presence or absence of blood in the spinal fluid will permit a ready distinction between the two. The value of angiography in identifying and localizing the lesion has become widely recognized, although here, too, limitations are imposed which, if ignored, will lead to false deductions.

The immediate causes of apoplexy fall readily into two categories: Vascular occlusion, and vascular rupture. Of these, occlusion will be considered first. Vessels leading to the brain or its subdivisions may be blocked by an embolus, by vascular spasm, or by a thrombus. Any of these may occur at any time and apparently in any circumstances, although thrombus is more liable to occur in a situation where the rate of blood flow is reduced.

The diagnosis of embolism is made when, associated with apoplexy, a potential source of embolus —such as cardiovascular thrombus, dislodged fat, or air—is recognized. Postpartum stroke also is felt to belong to this category.

In certain circumstances the onset of stroke from embolism may be heralded by a convulsive seizure. Involvement of consciousness may be associated with block of a large vessel or with interruption of certain of the smaller channels. Sensorimotor function is more commonly involved on the right side of the body, representing cerebral embolism on the left side, probably because of the mechanics of the vascular supply.

Spinal fluid pressure ordinarily is not elevated following embolic stroke, and in the acute stage the cell and chemical contents of the fluid are not materially altered. The block may be followed by extravasation of blood by diapedesis and later there may be frank rupture of the vessel due to the weakening of the vascular wall, but it is unlikely that these will play any major part in verifying the diagnosis of cerebral embolism. In the late stages, also, increase in the protein content of the cerebrospinal fluid may occur, particularly if an extended area of the brain surface has been affected.

Embolic apoplexy offers one of the most favorable situations for the therapeutic use of the anes-

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thetization of the cervical sympathetic chain, so-called "stellate block." Many times the patient is already under direct medical care because of the situation providing the source of the embolus; hence immediate treatment is possible with the result that damage to the cortical cells is minimal. Oftentimes the initial sympathetic block is all that is necessary, and if this is carried out within the first few minutes of onset, the return of function may be dramatic. Generally the use of niacin in a dose of 100 mg., from one to three times a day for several days after the stroke, seems desirable.

Vascular spasm may be confused with cerebral embolism as the cause of apoplectic ictus. Usually this is associated with other peripheral vascular disease, particularly arterial hypertension. In the young hypertensive person the appearance of vasospastic phenomena of itself indicates a vascular lability later eliminated by the sclerosing effect of degenerative disease. The vasospastic process may mimic embolism or thrombosis; it is recognized, of course, that vascular spasm is a significant factor in embolism, thrombosis and hemorrhage. The isolated effect of spasm alone seldom produces complete occlusion; the degree of effect, therefore, is usually not as great as when other factors are involved. This state of incomplete interruption of the blood flow permits of more latitude in time than with thrombosis or embolism.

In stroke due to uncomplicated vasospasm, the cerebrospinal fluid pressure, cell count and chemical contents are at first usually within normal limits. The onset of the disease is usually not quite so abrupt as with embolus or hemorrhage. Again the patient is often already under supervision and the onset of ictus immediately brought to the attention of a physician acquainted with the medical background. When cervical sympathetic block on the ipsilateral side is carried out early there is often rapid clinical evidence of the return of cerebral function, frequently dramatically manifested. Since the underlying process in vasospasm usually persists, recurrent episodes are not uncommon.

Thrombosis of cerebral vessels again presupposes vascular disease. Thrombosis may follow embolism or vasospasm and often occurs as a delayed manifestation in hemorrhage. Atheromatous plaques may provide the basis for the accumulation of clot. Interruption of the continuity of the endothelium is thought to be essential to the development of clot. It is well recognized that slowing of the rate of blood flow is a major contributing factor. Thrombosis producing apoplectic manifestations may occur in the vessels within the cranium itself or in the internal carotid artery distal to the bifurcation of the common carotid. Thrombosis in the cervical area is not uncommon and is usually followed by strikingly diffuse manifestations which vary to some degree depending upon the amount of collateral supply from other vessels contributing to the circle of Willis. The apposition of the internal carotid artery to the lateral surface of the atlas may result in demonstrable constriction of the artery, slowing the blood flow and occasionally producing thrombus at this point. Thrombosis in the basilar artery never, and in the vertebral artery seldom, is compatible with life.

Thrombosis, when recognized as an acute problem, may be treated with some success in the manner that is suggested for embolism and vasospasm. In a considerable number of cases the problem is one of chronic paralysis, probably the result of thrombosis. Occasionally the differentiation between thrombosis and hemorrhage will be difficult in these chronic situations. In such circumstances cerebral angiography is strongly advocated. In angiographic studies, angioma and aneurysm have been encountered in some cases, and extensive thrombotic involvement of one or both internal carotid arteries also has been observed. The information thus obtained is most valuable in advising the patient as to therapy and in estimating prognosis.

The treatment of chronic effects of thrombosis produces a less satisfactory result than that which may reasonably be anticipated in the acute stage. Many of the patients have profound emotional as well as sensory and motor afflictions, the former representing extensive basal ganglion involvement. It is necessary for both physician and patient to recognize at the outset that return to completely normal function is unlikely. Interruption of the cervical sympathetic nerve by procaine has been tried. Electroencephalograms taken immediately before and after the block, and careful objective and subjective analysis of change in the sensorimotor or emotional spheres, provide the basis of evaluation. If the result of chemical nerve block seems adequate, cervical sympathectomy or stellate ganglion decentralization may be indicated. Repeated stellate block will be disappointing, for the effectiveness of the local anesthetic agent has been observed to diminish rapidly. Intravenous histamine injections, suggested by Furmanski,[1] have been of value in instances where a surgical procedure does not seem justified. The administration of 1 mg. of histamine in 250 cc. of Ringer's solution or 5 per cent dextrose solution, intravenously, at a rate sufficient to produce a flush, and repeated as often as three times a week, has been followed by significant somatic and emotional improvement.

Embolism, vasospasm and thrombosis are all secondary manifestations of some underlying disease in the body. The necessity of treating the underlying disease must, of course, always be remembered and the cerebral problem regarded in its true situation as only one aspect of a general medical problem.

Apoplexy due to hemorrhage is the other broad division of this general problem. In young adults, cerebral hemorrhage may often be the result of rupture of a congenital saccular aneurysm or the result of rupture of vessels in an arteriovenous angioma. Hemorrhage may also result from vascular tear at the site of an arteriosclerotic plaque, from arteriovenous fistula, or from degenerative change in a tumor.

Hemorrhagic stroke in young adults can occur at any time. A demonstrable association between the onset and some physical or emotional strain is not consistently observed. It is reasonable to assume that rupture of a weak spot in the vascular tree is more liable to occur when factors which elevate the blood pressure come into play. However, hemorrhage is known to have occurred while the patient was engaged in conversation, while shaving, while lying in bed, after awakening in the morning, and while indulging in many other of the normal daily activities of life not generally associated with a particular alteration in blood pressure.

The hemorrhagic aspect of an ictus is generally diagnosed by the headache progressing to pain and stiffness in the cervical spine or by the overwhelming interruption of all conscious cerebral activity. Usually it can be proved by the demonstration of blood in the cerebrospinal fluid after such an incident. It is possible, however, for intracranial bleeding to occur without immediate significant change in the cerebrospinal fluid. Spinal puncture to establish the diagnosis of brain hemorrhage is felt to be unattended by any significant increase in hazard by reason of possible dislodgement of clot.

Cranial bruit is sometimes heard following hemorrhage, and may indicate an arteriovenous fistula or arteriovenous angioma. On the other hand, hemorrhage into the cranial cavity may distort the vascular tree to such an extent that bruit may result. The presence of bruit is not consistently helpful in determining the site of hemorrhage or even the side on which it occurs. Nor can the vessel bleeding be identified consistently by observation of clinical symptoms preponderantly referable to one side or the other, for aneurysms of the circle of Willis rupturing on its medial aspect may produce their major destruction in the contralateral hemisphere. Hemorrhages from the anterior communicating artery notoriously produce clinical symptoms on either side or even symptoms that suggest a rupture in the posterior part of the head.

The immediate treatment after hemorrhagic stroke is purely supportive in an attempt to permit the brain to adjust to the sudden accumulation of intracranial blood and the clotting mechanism to stem the flow from the tear in the vessel. Indiscriminate ligation of vessels in the neck on clinical grounds alone cannot usually be defended. Bleeding of hypertensive patients to reduce the arterial pressure has theoretical advantages, although the demonstrable evidence of help from this maneuver is open to some question. Cranial decompressive procedures in the acute stage are seldom if ever of sufficient value to justify the additional strain of a surgical procedure carried out at a time of crisis.

Direct and positive approach to the treatment of the bleeding point necessarily rests entirely upon the demonstration of the location and character of the bleeding point by cranial angiography. Angiograms should be obtained as soon after hemorrhage as the patient's general condition permits. Angiography should never be attempted, however, when the patient's condition is precarious. On the other hand, there is as yet no evidence that an injection of Diodrast® or Thorotrast® into the common carotid artery through a needle up to 18 gauge in size will materially increase the pressure in that artery. No rise in pressure was noted in direct readings in a small number of cases.* Actually, in some instances there was an apparent fall in pressure which theoretically might be related to the slowing of the pulse so often attendant upon injection of Diodrast.

Angiograms may demonstrate aneurysm or angioma, or may show no abnormality. For aneurysm arising from the carotid artery itself, ligature in the neck is the preferred treatment. This is usually done under local anesthesia and is sometimes followed by intracranial trapping. A pedunculated saccular aneurysm may be clipped across the neck with a good chance of producing cure. An atheromatous plaque at the base may prevent the accomplishment of this endeavor, however, and such plaques are not demonstrable except by actual observation in the operating room.

For aneurysms distal to the bifurcation of the internal carotid artery into the anterior and middle cerebral arteries, direct approach is necessary — either ligation of the entire vessel upon which the aneurysm arises, or, preferably, if it is possible, ligation across the base of the aneurysm so that the continuity of the supplying vessel is preserved. Aneurysms of the anterior communicating artery are particularly difficult to handle because they frequently receive major supply from both anterior cerebral vessels. The interruption of both is not consistent with life. Some investigators believe that aneurysms in this region should be completely abandoned, and the patient's fate left entirely to chance. In certain instances the aneurysms definitely are not operable; on the other hand, the establishment of a fixed ruling seems unreasonably defeatist.

Arteriovenous fistula has been found to exist between the posterior cerebral artery and the vein of Galen, and between the carotid artery and the cavernous sinus. Often the contributing artery can be interrupted and when this is the internal carotid the procedure is more safely done under local anesthesia. Intracranial trapping, however, may be necessary to achieve adequate block of the fistula.

Arteriovenous angioma, a completely inoperable lesion prior to the development of cranial angiography, is now found to be operable in about 50 per cent of instances. By means of angiography the major contributing vessel can usually be demonstrated. and with experience the auxiliary contributors from the cerebral depths can be anticipated and controlled. Deep-lying angiomas involving the entire thalamus, or smaller ones which have arisen in the vicinity of the vein of Galen, have generally been avoided unless by repeated hemorrhage the excessive danger of the lesion to life becomes evi-

* The readings were taken by Dr. Lawrence Arnstein in the neurological surgery laboratory of the University of California School of Medicine.

dent. Those tumors whose vessels are at all accessible have been operated upon, the purpose being to prevent the patient's dying from massive hemorrhage. Such an end can be anticipated if the angioma is not so approached: In the author's experience a considerable proportion of patients with angiomas that were not operated upon died during the period of observation, usually within a matter of one to three years.

For the third group of patients, those who have had objective evidence of hemorrhage but in whom the arteriogram demonstrates no source of bleeding, sharp limitation of activity with absolute bed rest for at least two months and gradual return to activity approaching normal for the next two months, is recommended. Failure of demonstration of the source of bleeding has been interpreted by the author to mean that the bleeding was from a small tear, as at an atheromatous plaque, or that it was from an aneurysm which had become filled with clot between the time of hemorrhage and the time of angiography. The rationale of absolute bed rest for two months is to give adequate time for the clot to become replaced by scar tissue, which, hypothetically, will be sufficiently firm in that time to justify the gradual reestablishment of normal activity. The patient always is warned against physical or emotional strain on the hypothetical grounds mentioned earlier. This mode of treatment is an adaptation of the so-called conservative treatment which previously was recommended in all cases of cerebral hemorrhage. It has been successful in treatment of patients without demonstrable lesions. But in the author's experience it has not been successful for patients with a demonstrable lesion; in several instances the patients died, some of them during the period of prolonged bed rest.

In evaluating the results of treatment in apoplexy in young adults, it is necessary to recognize first the danger of the underlying disease process, and secondly the probable limitations of any therapeutic measure. In occlusive apoplexy the danger to life is usually greatest at the time of or immediately following the occlusion. The benefit from vasodilating mechanisms is directly proportional to the promptness with which they can be instituted and to the size and location of the vessel affected. In general, vessels mainly supplying the brain cortex react favorably to treatment by vasodilating mechanisms. If the degenerative change is in the white matter or tracts of the brain, the evidence is that no improvement can be anticipated.

In the matter of hemorrhagic apoplexy the danger to life exists until the source of bleeding is controlled or isolated from the circulation. The primary end, then, is preservation of life. A secondary consideration is the limitation of the degree of residual morbidity. Frequently saccular aneurysm, and occasionally arteriovenous angioma, may be adequately dealt with without apparent morbidity. Some morbidity is often associated with the ictus itself where the source is aneurysm, and this can usually be anticipated with angioma. Vasodilating mechanisms may minimize the morbidity from a hemorrhagic ictus to the degree that auxiliary vasospastic phenomena have affected the adjacent cortex. Much hemorrhagic damage is to the white matter, however, and here again no benefit can be anticipated.

The problem of morbidity and mortality must in some way be brought to the consideration of the patient, for in the last analysis he must make the decision. In the author's experience, most patients prefer some incapacity to a continued existence under a Damoclesian sword.

REFERENCE

1. Furmanski, A. R.: Histamine therapy in acute ischemia of the brain, Arch. Neurol. & Psychiat., 63:415-424, March 1950.